Intramedullary Solitary Fibrous Tumor—A Benign Form of Hemangiopericytoma? Case Report and Review of the Literature

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Key words
- Hemangiopericytoma
- Intramedullary
- Neurosurgery
- Solitary fibrous tumor
- Spinal cord

Abbreviations and Acronyms
HPC: Hemangiopericytoma
MRI: Magnetic resonance imaging
SFT: Solitary fibrous tumor

INTRODUCTION

Solitary fibrous tumors (SFTs) are mesenchymal tumors first described in 1931 by Klemperer and Rabin as a primary neoplasm arising from the pleura (23). The visceral pleura is still the predominant location of SFTs. Nevertheless, SFTs can arise in the soft tissue anywhere in the body, even in the central nervous system (2, 7, 8, 15, 18, 21, 24, 32, 41-43). The occurrence completely or partially within the spinal cord is rare and has been reported in 16 cases so far to the best of our knowledge (1, 4, 8, 10, 11, 17, 18, 20, 21, 27, 29, 33, 34, 38).

Although there are some recurrent cases even after complete resection (3, 4, 11, 21, 27, 38), SFTs of the central nervous system are principally benign tumors that can be treated by surgery alone, as they seem to be insensitive to chemotherapy or radiotherapy (5, 6, 28, 31). However, the histological findings of SFT show many similarities to the more malignant hemangiopericytoma (HPC), which is also rare in the central nervous system (5, 10, 36, 38). In the past decade more and more pathologists assume a common spectrum of these tumors (5, 28).

We present a case of an intramedullary tumor with typical histopathological and immunohistochemical findings for SFT.

CASE DESCRIPTION

An 83-year-old female presented with a 1.5-year history of weakness and progressive paresthesia of the left leg. Recent to admission the symptoms had worsened to bilateral spastic paralysis of both legs and bladder incontinence. Further examination revealed 3/5 paresis of the legs, and even with assistance the patient was unable to walk due to uncontrollable spasticity and ataxia of both legs. Sensory testing revealed numbness of the lower left leg. Magnetic resonance imaging (MRI) showed an intraspinal tumor at the level of T8-T9 with a massive compression of the spinal cord. The lesion was slightly hyperintense in T1-weighted images and hypointense in the T2-weighted images and showed a homogeneous gadolinium contrast enhancement (Figure 1). The lesion was suspected to be a meningioma, and an operation was performed rather quickly in order to restore the neurological function.

The operation was carried out through a laminectomy of T9 and a partial laminectomy of T8 and T10. An intradural, mainly intramedullary lesion was exposed as the tumor had been suspected to be the tumor with typical histopathological and immunohistochemical findings for SFT.

CASE REPORT

The lesion with typical histopathological and immunohistochemical findings for SFT.

BACKGROUND: Solitary fibrous tumors (SFTs) are benign tumors of the soft tissue occurring anywhere in the human body but arise predominantly in the visceral pleura. SFTs of the central nervous system are rare, especially when they occur within the spinal cord.

CONCLUSIONS: There is evidence that SFTs and hemangiopericytomas (HPCs) are not different entities but should be considered as different gradations of a common spectrum. The extent of resection is a prognostic factor for recurrence-free survival in SFT; therefore we recommend surgery with complete resection whenever possible depending on the results of mandatory intraoperative neurophysiological monitoring in these cases.
the remaining tumor part 9 days later under neurophysiological monitoring conditions. The tumor was well circumscribed but showed some arachnoid invasion on the caudal and medial parts. With the microsurgical technique complete, tumor removal was achieved.

The neurological function improved slowly after the operation, and the patient was sent to a rehabilitation center. At that time she was able to stand without assistance. Eight months later motor function of the legs has improved to 4/5 according to Medical Research Council (MRC) motor grading. She is able to walk a few steps but is dependent on a wheelchair for longer distances due to persistent ataxia.

Histological Findings

Eight fragments of unfixed tissue obtained within the first operation were transferred to the neuropathology department. Fresh frozen, hematoxylin-eosin stained sections of some fragments revealed a relatively dense tumor tissue with spindle-shaped tumor cells arranged in fascicles. The nuclei were oval to elongated lacking increased mitotic activity. There was no necrosis.

Remaining parts of the tissue sample were fixed in formalin and embedded in paraffin (Figure 2). Tissue sections showed again a spindle cell tumor with intermingled collagen fiber bundles and a dense reticulin fiber network. Mitoses were rare with less than 1 in 10 high-power field (HPF) and no necrosis. Immunohistochemical analysis revealed strong expression of vimentin but was negative for S100 and epithelial membrane antigen (EMA). The tumor displayed a strong nuclear STAT6 staining. There was a patchy immunoreactivity for bcl-2 and CD34. MIB1-labeling was low in most parts of the tissue samples, mostly not exceeding 1%; however, focally the labeling-index increased up to 5%. On the basis of the morphologic and immunohistochemical findings, the tumor was diagnosed as an SFT.

The tissue specimen from the second surgery revealed again a spindle cell tumor rich in collagenous connective tissue (Figure 3). Still, mitoses were rare. There was focal necrosis, which we attribute to the first surgery. Furthermore, this tissue specimen displayed large areas of collagen depositions with fibrosis and hyalinization with focal remnants of peripheral nerves, which seemed to be diffusely invaded by the tumor. The immunohistochemical expression pattern corresponded to the tumor tissue of the first surgery and confirmed the diagnosis of an SFT.

DISCUSSION

SFT is a rare, predominantly benign lesion that can arise anywhere in the soft tissue of the human body. Nevertheless, recurrence is known and malignant behavior of SFT involving the central nervous system has been reported in a few cases (5, 6, 8, 11, 13, 14, 30, 31, 38, 39).

The majority of SFTs of the central nervous system are intracranial, intradural, extraxial lesions (3). More than a third (34%) occur within the spinal canal, and nearly half of these (49%) are located partially or completely intramedullar (27). However, intramedullary SFTs are still a rare entity. To the best of our knowledge, only 17 cases including the present one have been published so far (1, 4, 8, 10, 11, 17, 18, 20, 21, 27, 29, 33, 34, 38).

SFTs of the spinal canal are most often localized in the thoracic spine followed by the cervical spine (11, 18). Of the 17 cases with intramedullary components, half were located cervical and the other half in the thoracic region. The reviewed cases of intramedullary SFT showed no adherence to the dura mater but strong adherence to the pia mater or spinal cord. In these cases the origin is suspected to be the pia mater itself or the perivascular soft tissue (17, 18). All reviewed intramedullary cases were treated surgically, and complete removal was achieved in 79% (Table 1).

Despite the fact that SFTs are usually isointense or hypointense in T1-weighted images, the lesion in the present case was slightly hyperintense in T1-weighted images. However, histological examination revealed characteristic findings for SFTs. Signal intensity in T2-weighted images is typically hypointense due to hypercellular
areas or hypocellular but collagenous or calcified areas within the tumor \((11, 12, 18, 21, 22, 26, 27, 37)\). SFTs show a homogeneous gadolinium enhancement in nearly every case (see Table 1). These MRI features make it difficult to distinguish SFTs from meningioma, schwannoma, and other intradural and even intramedullary tumor entities like astrocytoma or ependymoma \((3, 11, 17, 18, 21, 27, 31)\). Because of MRI, the initial diagnosis in the presented case was meningioma, although intraoperative findings revealed the main part of the tumor to be located intramedullary without dural attachment. This misleading preoperative assumption is known for SFTs even with intramedullary parts \((1, 27, 38)\).

In 23\% of the reviewed intramedullar cases the lesion was preoperatively suspected to be extramedullar, and in another 23\% of cases discrimination between intramedullary and extramedullary was not possible by MRI (see Table 1).

SFT is a spindle cell tumor characterized by a combination of alternating hypocellular collagenic and hypercellular areas showing branching vessels \((5)\) with similarities to fibrous meningioma \((19)\) and hemangiopericytoma \((40)\). SFTs are typically positive for CD34 and can be distinguished from schwannoma and meningioma via negative reaction to S100 and EMA in immunohistochemistry \((5, 11, 18, 21, 27)\). Furthermore, SFTs are positive for vimentin \((11, 18, 21)\) and bcl-2 \((9, 11, 27)\). MIB-1 labeling is low \((3, 5, 18)\).

It is difficult to distinguish SFTs from HPCs via MRI, histology, or immunohistochemistry as both show many overlapping features \((5, 10, 38)\). Even though HPC is a rare tumor of the central nervous system, intramedullary occurrence has been described in a few cases \((36)\). In contrast to SFTs, HPCs are typically hypercellular with a higher mitotic rate and presence of cell atypia, show CD34 positivity in 33\%–100\%, and are highly vascularized \((5, 6, 16, 18, 25, 38)\). A cellular form of SFT that is indistinguishable from HPC has been described. In the past decade many pathologists have proposed a
### Table 1. Cases of Intramedullary Solitary Fibrous Tumor in Literature

<table>
<thead>
<tr>
<th>Author</th>
<th>Sex/Age</th>
<th>Loc.</th>
<th>Symptoms</th>
<th>MRI Findings</th>
<th>Intraoperative Findings</th>
<th>Follow-up</th>
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<td>Def. Location</td>
<td>EoR</td>
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<td>Treatment</td>
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<td>Spinal cord</td>
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<td></td>
<td>Neurological</td>
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<td>Neurol. outcome</td>
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<td></td>
<td>Recurrence</td>
<td></td>
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<tr>
<td>Carneiro et al. 1996</td>
<td>m/50</td>
<td>-</td>
<td>Cono-caudal syndrome</td>
<td>-</td>
<td>Surgery</td>
<td>IM + EM</td>
</tr>
<tr>
<td>Alston et al. 1997</td>
<td>m/47</td>
<td>T 4-5</td>
<td>Brown–Ségard syndrome</td>
<td>-</td>
<td>Surgery</td>
<td>IM</td>
</tr>
<tr>
<td>Kanahara et al. 1999</td>
<td>m/62</td>
<td>C 6-7</td>
<td>Leg numbness</td>
<td>Hypo</td>
<td>Surgery</td>
<td>IM + EM</td>
</tr>
<tr>
<td>Mordani et al. 2000</td>
<td>m/33</td>
<td>C 5</td>
<td>Cervical myelopathy</td>
<td>Iso</td>
<td>Surgery</td>
<td>IM</td>
</tr>
<tr>
<td>Tihan et al. 2003</td>
<td>m/50</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>Surgery</td>
<td>IM</td>
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<td>Tihan et al. 2003</td>
<td>m/50</td>
<td>-</td>
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<td>Hypo</td>
<td>Surgery</td>
<td>IM</td>
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<tr>
<td>Kawamura et al. 2004</td>
<td>m/64</td>
<td>T 2-3</td>
<td>Brown–Ségard syndrome</td>
<td>Hypo</td>
<td>Surgery</td>
<td>IM + EM</td>
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<tr>
<td>Bohinski et al. 2004</td>
<td>f/49</td>
<td>C 4</td>
<td>Neck pain, arm paresthesia</td>
<td>Iso</td>
<td>Surgery</td>
<td>IM + EM</td>
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<tr>
<td>Jallo et al. 2005</td>
<td>m/41</td>
<td>C 8-7</td>
<td>Cervical myelopathy</td>
<td>Hypo</td>
<td>Surgery</td>
<td>IM + EM</td>
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<tr>
<td>Jallo et al. 2005</td>
<td>m/17</td>
<td>T 5-6</td>
<td>Spastic paraparesis</td>
<td>Hypo</td>
<td>Surgery</td>
<td>IM</td>
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<tr>
<td>Pizzolitto et al. 2005</td>
<td>m/47</td>
<td>C 4</td>
<td>Cervical myelopathy</td>
<td>-</td>
<td>Surgery</td>
<td>IM</td>
</tr>
<tr>
<td>Ishii et al. 2009</td>
<td>f/63</td>
<td>C 5</td>
<td>Weakness upper extremity</td>
<td>Iso</td>
<td>Surgery</td>
<td>IM</td>
</tr>
<tr>
<td>Cappetta et al. 2010</td>
<td>f/75</td>
<td>T 6-7</td>
<td>Paralysis lower extremity</td>
<td>Hypo</td>
<td>Surgery</td>
<td>IM + EM</td>
</tr>
<tr>
<td>Fargen et al. 2011</td>
<td>f/28</td>
<td>C 2-3</td>
<td>Lower extremity numbness</td>
<td>-</td>
<td>Surgery</td>
<td>IM + EM</td>
</tr>
<tr>
<td>Mariniello et al. 2012</td>
<td>m/75</td>
<td>T 6-7</td>
<td>Thoracic myelopathy</td>
<td>Hypo</td>
<td>Surgery</td>
<td>IM</td>
</tr>
<tr>
<td>Robert et al. 2014</td>
<td>f/49</td>
<td>T 9-10</td>
<td>Thoracic myelopathy</td>
<td>Hypo</td>
<td>Surgery</td>
<td>IM + EM</td>
</tr>
<tr>
<td>Present case</td>
<td>f/83</td>
<td>T 8-9</td>
<td>Paralysis and incontinence</td>
<td>Hyper</td>
<td>Surgery</td>
<td>IM + EM</td>
</tr>
</tbody>
</table>

m, male; f, female; T, thoracic; C, cervical; hypo, hypointense; iso, isointense; homo, homogeneous; IM, intramedullar; EM, extramedullar; NL, neurological; Loc., Location; Susp., suspected; Def., definitive; EoR, extent of resection; attachm., attachment; Mths., months; Rec., recurrence; Neurol., neurological.
common spectrum of these tumors (5, 28). Because SFT progression to HPC-like tumors and HPC recurrence as SFTs have been reported, Bouvier et al. (5) suggested that the artificial separation between SFT and HPC is a reflection of histological appearance and does not indicate two distinct entities. This suggestion was strongly supported by the findings of Schweizer et al. (35), who reported a similar NAB2-STAT6 fusion in the DNA of HPC and SFT.

Some authors have already recommended clinical treatment and follow-up strategies according to HPC treatment (6). However, SFTs are predominantly benign. Anaplastic HPCs are malignant neoplasms with higher recurrence rates, with a tendency for meningeval spreading, and distant metastases (5, 38) necessitating a more radical treatment.

The extent of resection is a prognostic factor for recurrence-free survival in SFT, and surgery is recommended. Complete removal of the tumor is possible even in SFT with intramedullary components. Resection should be performed carefully under neurophysiological monitoring because most SFTs are well defined (see Table 1) but can present with a firm spinal cord attachment (5, 27). Thereby a favorable outcome is achievable. Recurrence is rare after gross total removal but has been reported after subtotal resection despite subsequent radiotherapy (3, 4, 11, 21, 27, 38). Recurrence of SFT of the central nervous system can occur several years after clinical manifestation (8, 30, 38, 40), and drop metastasis after complete resection has been described (34).

Chemotherapy has been administered in some recurrent cases, but so far there is no evidence of a therapeutic effect (6). SFTs seem to be insensitive for radiotherapy and chemotherapy (5, 28, 31).

As other authors have already proposed (10, 13, 17, 28), we suggest a careful clinical and radiological long-term follow-up in cases of SFT of the central nervous system.

CONCLUSION

Intramedullary SFTs are rare entities with predominantly benign behavior. Consid- ering histopathological examination, SFTs and HPCs seem to be different gradua- tions of the same tumor entity. The extent of resection is a prognostic factor for recurrence-free survival. Therefore surgery is recommended and intraoperative neurophysiological monitoring is manda- tory in intramedullary cases. Complete resection should be aspired whenever possible depending on monitoring results.

REFERENCES


